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CASE-BASED NEURO-OPHTHALMOLOGY

CONTENTS

CHAPTER 11.

Case

Age: 55-year-old black man

Reason for referral to ophthalmology: Glaucoma

Past medical history: None

Past ocular history: None

Medications: None

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Next: Pituitary apoplexy

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Family history: Father with glaucoma treated with topical medications

HPI: He went to see an optometrist for a routine examination and was found to have optic nerve cupping in both eyes. His intraocular pressure was 20 OD and 21 OS and his central corneal thickness was 550 μm OD and 546μm OS. An ophthalmology consultation was requested for further management.

Ophthalmological examination:

Blood pressure: 118/80, heart rate 74

Visual acuity: 20/20 OD, 20/20 OS

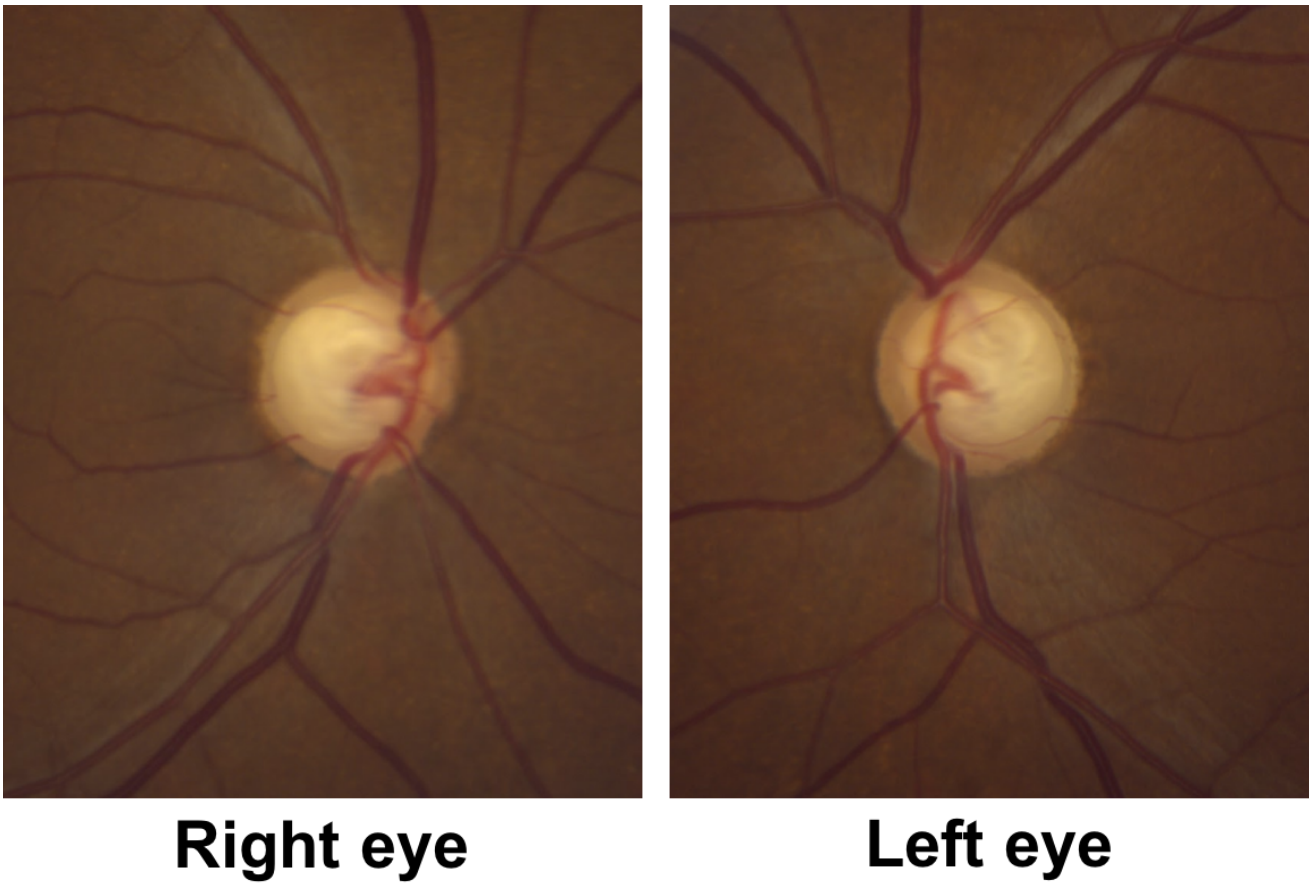
Pupils are equal sizes and reactive to light, there is no RAPD

Color vision: 14/14 OD and 14/14 OS correct Ishihara plates

Ocular motility and alignment are normal

Slit lamp examination is normal

Neurological examination is normal



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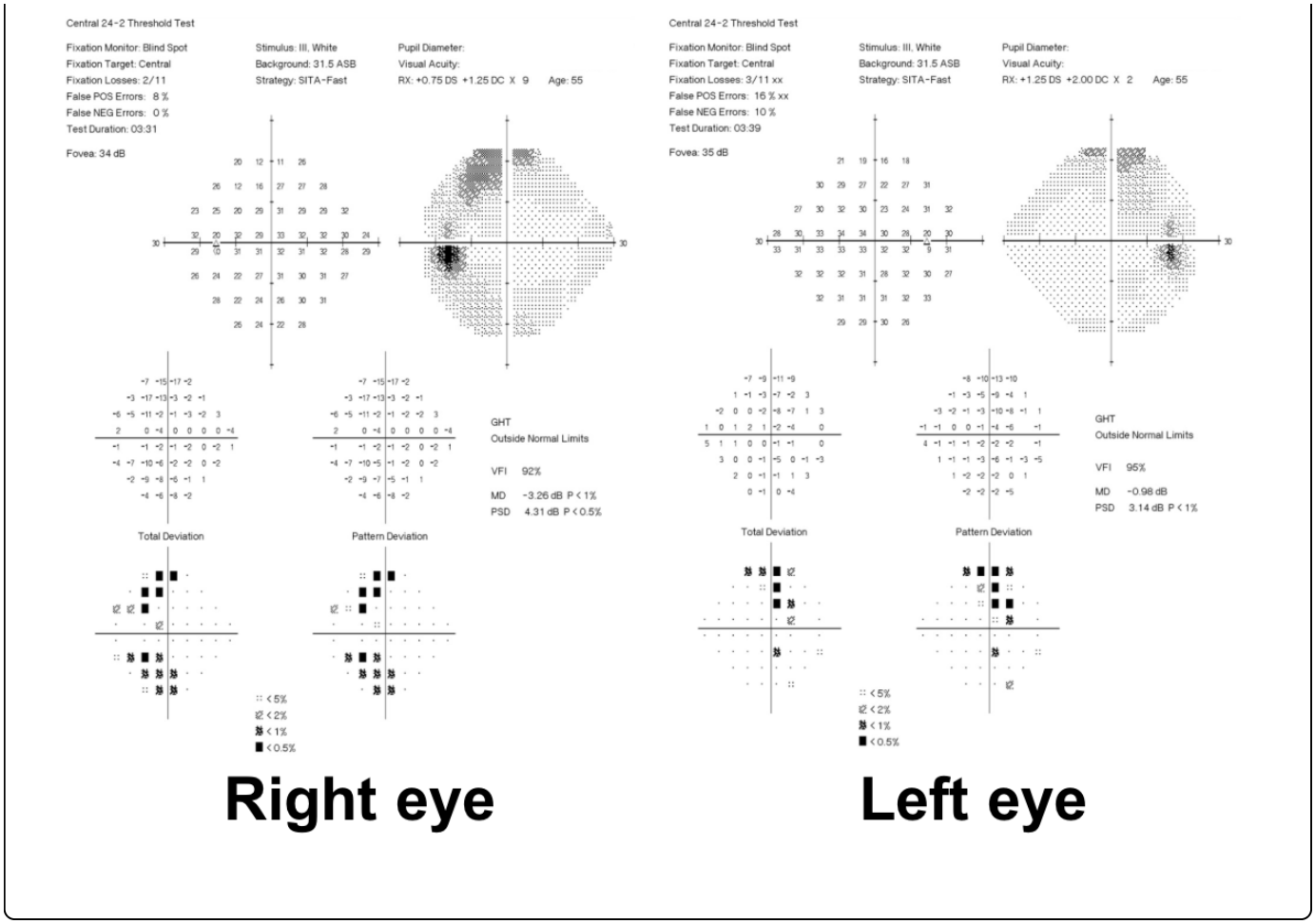

1. Which of the following is true regarding intraocular pressure (IOP) and glaucoma?

1. All patients with glaucoma have elevated IOP at presentation
2. IOP can vary by 10 mm Hg or more over a period of 24 hours in patients with glaucoma
3. IOP is an adequate screening test for glaucoma
4. The mean IOP in the population is 21 mm Hg

1. Which of the following is true regarding intraocular pressure (IOP) and glaucoma?
2. IOP can vary by 10 mm Hg or more over a period of 24 hours in patients with glaucoma

Elevated IOP is an important risk factor for primary open angle glaucoma. The mean IOP in the population is 15.5 mm Hg with a standard deviation of 2.6. This has led to the "normal" range of IOP of 10-21, which is 2 standard deviations from the mean. Many population-based studies studies have found that as many as 30 to 50% of individuals with glaucoma and visual field loss have initial screening IOPs less than 22 mm Hg. Intraocular pressure can vary considerably, by more 10 mm Hg in 24 hours, in patients with glaucoma. Some patients with glaucoma have IOPs that are consistently within the "normal" range, which is termed normal-tension glaucoma. Patients with optic nerve cupping and normal IOPs often pose the most difficulty in differentiating glaucoma-

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Clinical Pearl

Intraocular pressure within the normal range does not rule out glaucoma.

2. What visual acuity is expected in a patient with moderate glaucoma and no other ocular pathology?

1. Normal visual acuity
2. Mildly reduced visual acuity
3. Moderately reduced visual acuity
4. Severely reduced visual acuity

2. What visual acuity is expected in a patient with moderate glaucoma and no other ocular pathology? 1. Normal visual acuity

Patients with glaucoma, even in advanced stages, usually have preserved visual acuity and colour vision since glaucoma tends to spare the papillomacular bundle. This is a useful clue in differentiating glaucomatous and non-glaucomatous optic neuropathy. The latter tends to involve the papillomacular bundle early, causing reduced central vision.

Clinical Pearl

Reduced visual acuity (especially without total cupping) is concerning for non-glaucomatous optic neuropathy.

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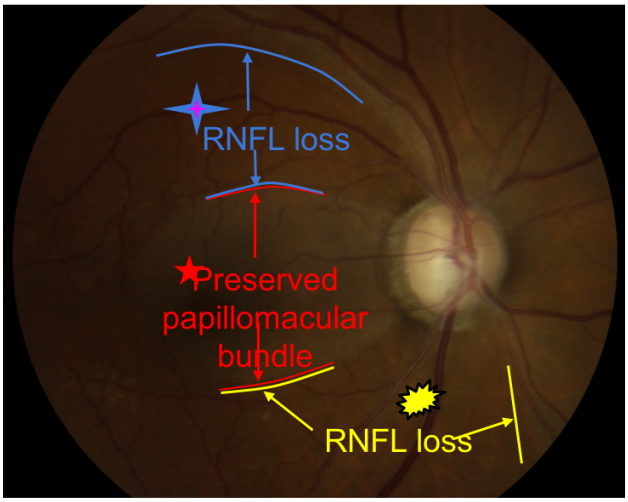

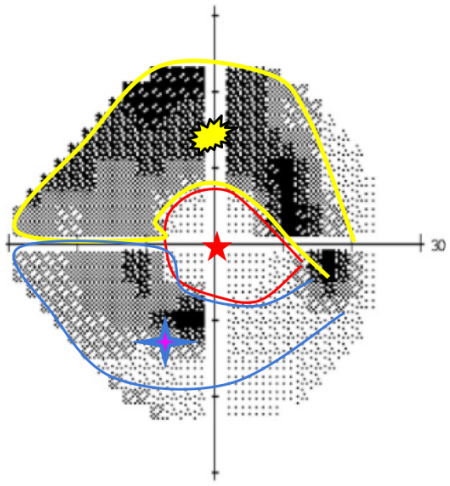

In this patient with primary open angle glaucoma, there is a superior arcuate defect corresponding to the inferior RNFL loss and inferior cupping (yellow) since the inferior retina is responsible for the superior visual field. There is a similar loss of superior RNFL corresponding to a milder inferior arcuate defect (blue). The papillomacular bundle, which is responsible for central and color vision is preserved (red).

3. What clues from the slit lamp examination would favour glaucoma over a non-glaucomatous optic neuropathy?

1. Pseudoexfoliative material on the lens capsule
2. Krukenberg spindle
3. Peripheral anterior synechiae
4. Narrow angles
5. All of the above

3. What clues from the slit lamp examination would favour glaucoma over a non-glaucomatous optic neuropathy? 5. All of the above

When assessing a patient with optic nerve cupping, signs of secondary glaucoma suggest a glaucomatous rather than a non-glaucomatous optic neuropathy. These signs include pseudoexfoliative material on the pupillary border or lens capsule (pseudoexfoliation syndrome), Krukenberg spindle, heavily pigmented trabecular meshwork, and iris transillumination defects (pigment dispersion syndrome), posterior or peripheral anterior synechiae and anterior chamber cells (uveitic glaucoma), and occludable or narrow angles (narrow angle glaucoma). Usually, these signs are associated with an elevated intraocular pressure.

4. Cupping occurs when there is:

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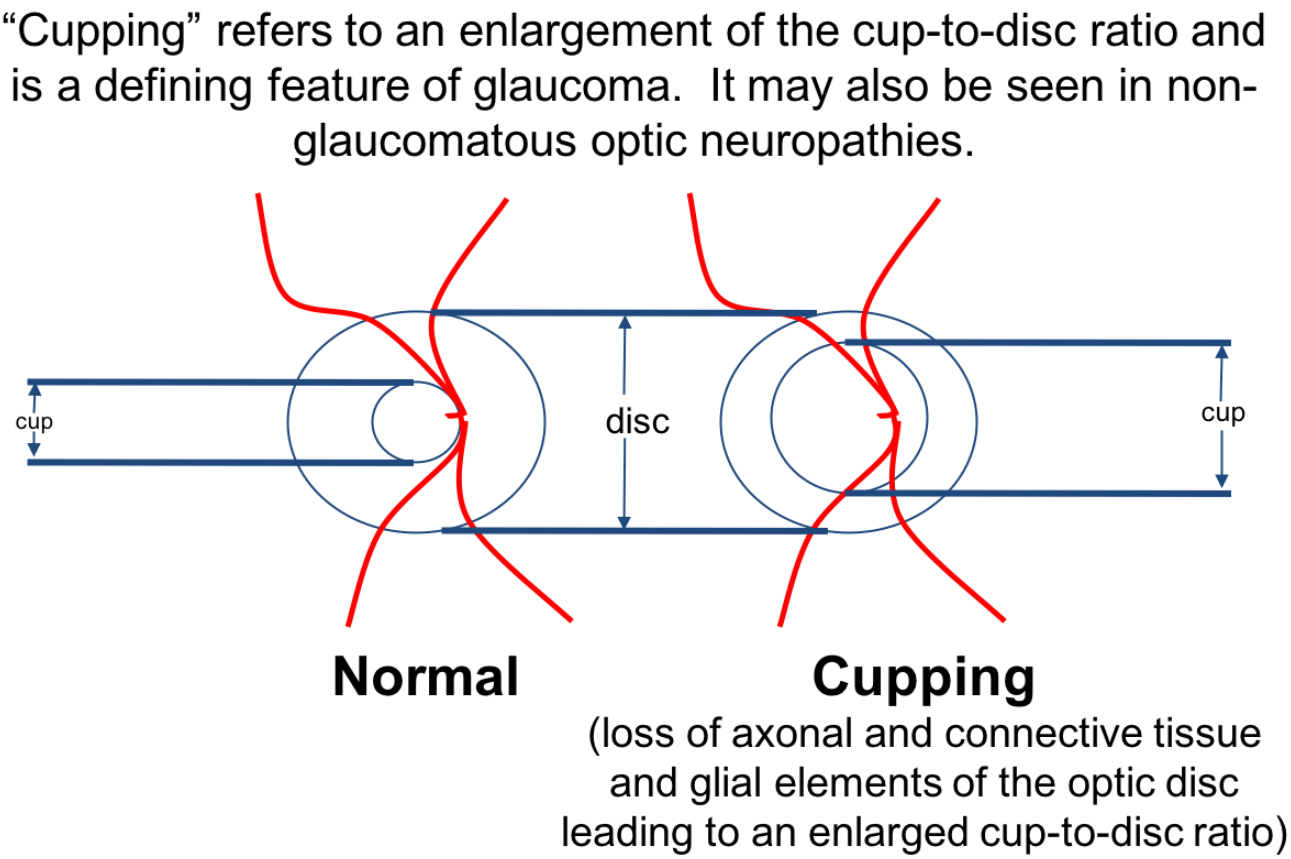
4. Choroidal thinning

4. Cupping occurs when there is: 2. Loss of retinal nerve fiber layer and connective tissue and glial elements of the optic disc

When there is loss of axons of the optic disc, but not the connective tissue and glial elements, the optic nerve appears pale and usually does not have significant cupping. When both the axonal and supporting connective tissue are lost, the nerve appears cupped as demonstrated below.


"Cupping" refers to an enlargement of the cup-to-disc ratio and is a defining feature of glaucoma. It may also be seen in non-glaucomatous optic neuropathies.
cup
disc
cup
Normal
Cupping
(loss of axonal and connective tissue and glial elements of the optic disc leading to an enlarged cup-to-disc ratio)

5. What visual field defect is expected in this patient with glaucoma?

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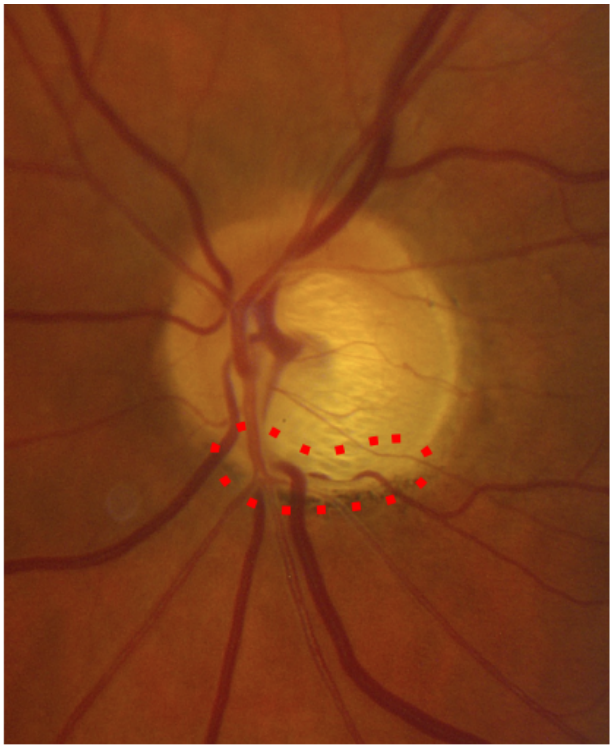
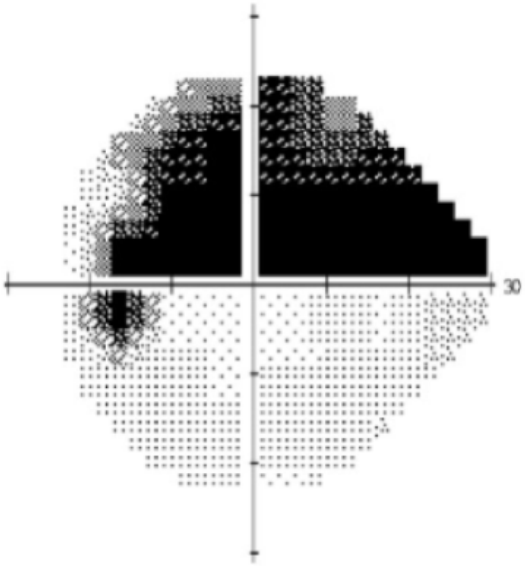
1. Severely constriction
2. Superior arcuate
3. Inferior arcuate
4. Central scotoma

5. What visual field defect is expected in this patient with glaucoma? 2. Superior arcuate

When assessing a patient for possible glaucoma, it is important to ensure that the optic nerve changes match the visual field defect. This patient has prominent loss of the neuroretinal rim inferiorly, which corresponds to a superior altitudinal visual field defect. If the optic nerve changes do not match the visual field or the visual loss is out of proportion to the optic nerve changes, than a non-glaucomatous optic neuropathy should be suspected.





Inferior thinning (red dotted circle) corresponds to a superior visual field defect since the inferior retina is responsible for the superior visual field.

6. Which of the following optic nerves most likely represents glaucoma?

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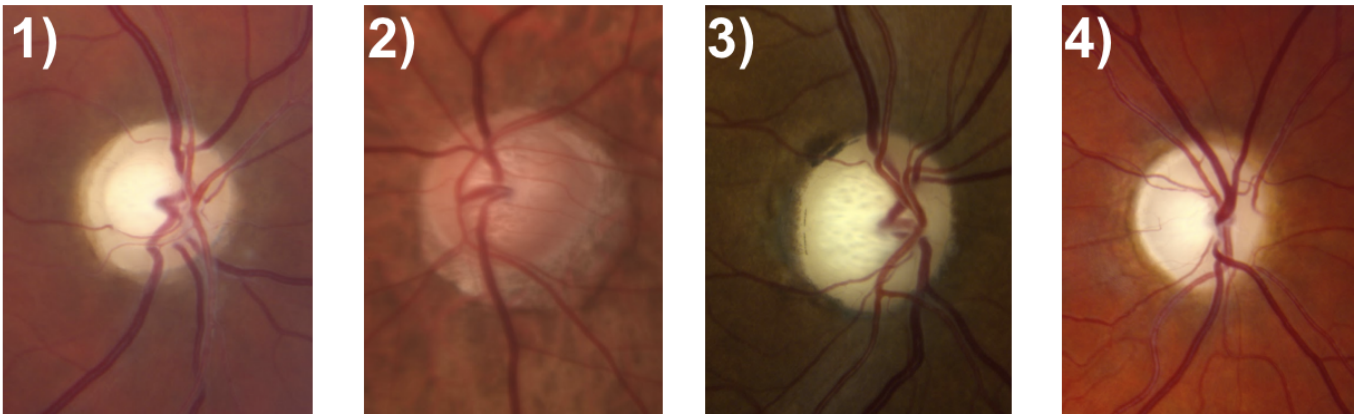
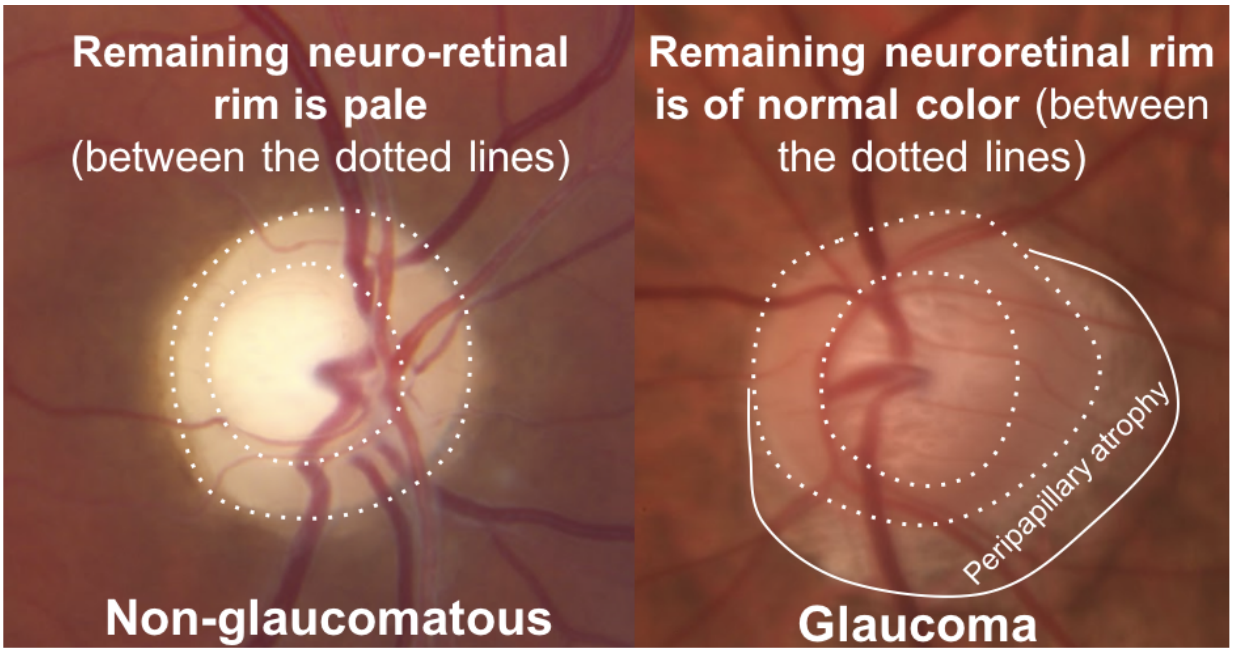


6. Which of the following optic nerves most likely represents glaucoma? 2.

The optic nerve depicted in (2) has an enlarged cup-to-disc ratio, but the remaining neuroretinal rim is a normal, pink color. The other optic nerves have pallor of the neuroretinal rim out of proportion to the cupping, which is characteristic of non-glaucomatous optic neuropathies. Other features of a glaucomatous optic nerve are listed in the box below.


Remaining neuro-retinal rim is pale (between the dotted lines)
Non-glaucomatous
Remaining neuroretinal rim is of normal color (between the dotted lines)
Peripapillary atrophy
Glaucoma

A comparison of a non-glaucomatous optic nerve with pallor and a glaucomatous optic nerve with normal color of the remaining neuro-retinal rim. Peripapillary atrophy is another sign suggestive of glaucoma.

It is not always possible to differentiate glaucomatous and non-glaucomatous optic neuropathies just by assessing the optic nerve. Optic nerve pallor is a subjective sign that is not always obvious and there may be little remaining neuroretinal rim to adequately assess this feature in patients with advanced disease. Some individuals may have anolmalous optic nerves and cupping cannot

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Clinical Pearl

Clinical features of a **glaucomatous optic nerve** include:

- Vertical elongation of the cup
- Focal elongation of the cup (notching)
- Normal color of the neuroretinal rim
- Excavation of the cup
- Peripapillary atrophy
- Disc hemorrhage (seen more frequently in normal tension glaucoma)
- Total cupping
- Exposed lamina cribrosa
- Baring of circumlinear vessels

7. Which optic neuropathies may demonstrate optic nerve cupping (enlarged cup-to-disc ratio)?

1. Compressive optic neuropathy
2. Hereditary optic neuropathy (Eg. Leber's Hereditary Optic Neuropathy)
3. Arteritic anterior ischemic optic neuropathy
4. Traumatic optic neuropathy
5. All of the above

7. Which optic neuropathies may demonstrate optic nerve cupping (enlarged cup-to-disc ratio)? 4. All of the above

Compression of the anterior visual pathways from tumors or aneurysms, hereditary optic neuropathies, arteritic ischemic optic neuropathies, and traumatic optic neuropathies may all cause cupping. Shown below is a patient with a right compressive optic neuropathy from an internal carotid artery aneurysm. Notice that there is cupping and pallor in the right eye, which is more obvious in comparison to the normal, left eye.

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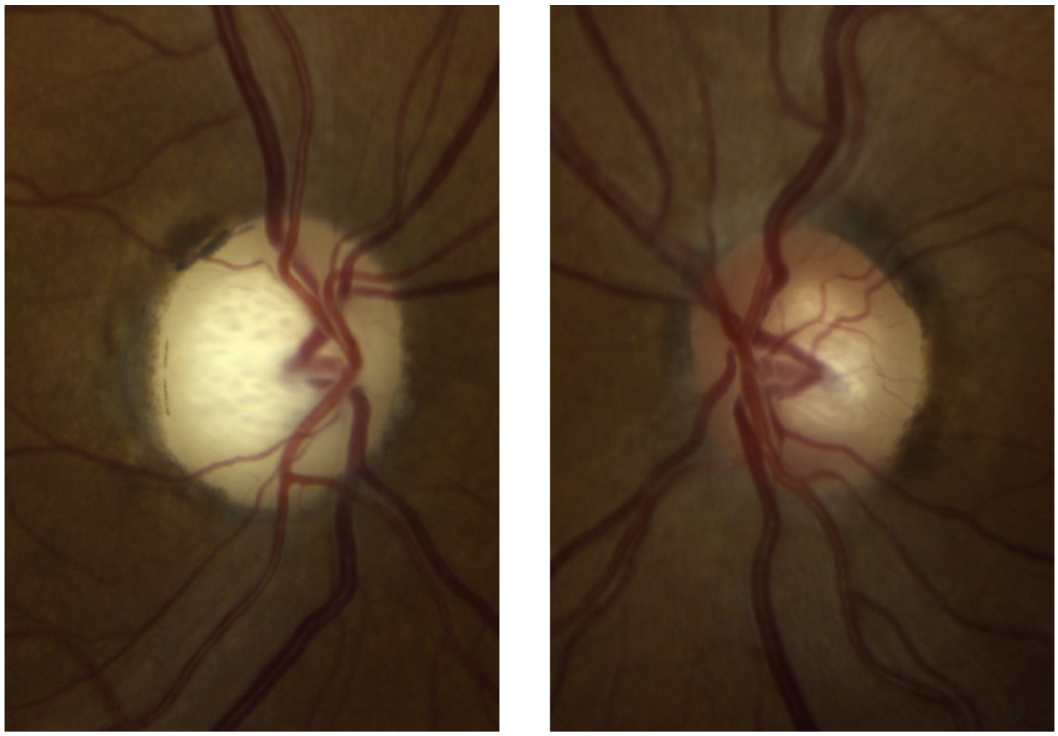

Right eye Left eye

There is an enlarged cup-to-disc ratio and pallor in the right eye compared to the left eye in this patient with a right compressive optic neuropathy secondary to an internal carotid artery aneurysm.

8. What other feature from the history or physical exam would be concerning for a non-glaucomatous optic neuropathy?

1. Rapid progression of vision loss
2. Intolerance of topical glaucoma medications
3. Family history of glaucoma
4. Previous use of longterm topical steroids

8. What other feature from the history or physical exam would be concerning for a non-glaucomatous optic neuropathy? 1. Rapid progression of vision loss

Although there are a number of factors that determine progression, glaucoma is typically a slowly progressive disease. A patient with rapidly progressing vision loss without an adequate explanation (such as uncontrolled IOP) should be assessed for non-glaucomatous etiologies.

9. Returning to the original case, what do the optic nerves (shown below) demonstrate?

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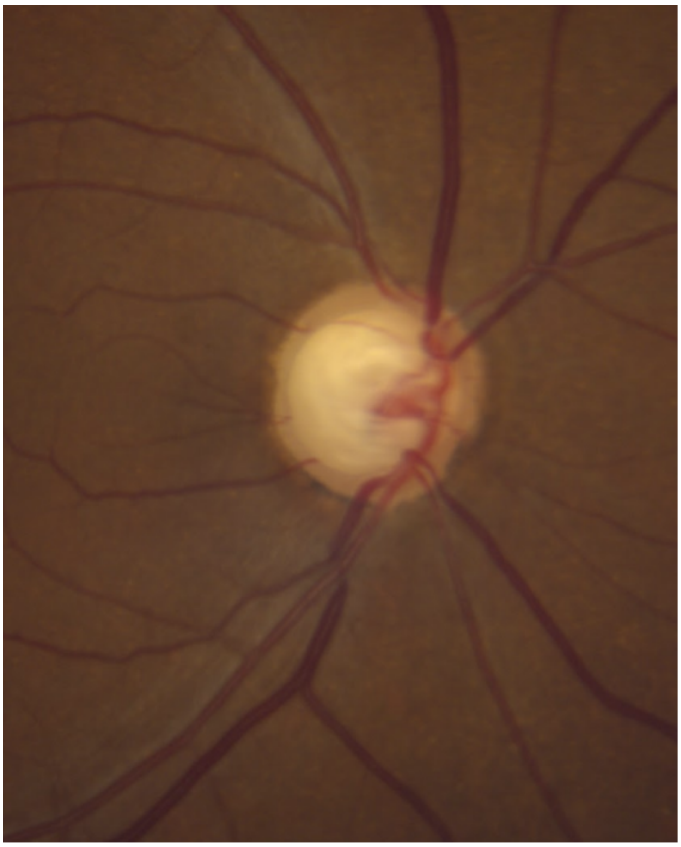

Right eye

Left eye

1. Bilateral optic nerve pits
2. Cupping with pallor of the remaining neuroretinal rim
3. Optic disc edema
4. Optic disc hemorrhages

9. Returning to the original case, what do the optic nerves demonstrate? 2. Cupping with pallor of the remaining neuroretinal rim

This patient has obvious cupping with a cup-to-disc ratio of approximately 0.9. There is also pallor of the remaining neuroretinal rim, although this is not as obvious as the non-glaucomatous optic nerves shown in question 6.

10. What visual field defect does this patient have?

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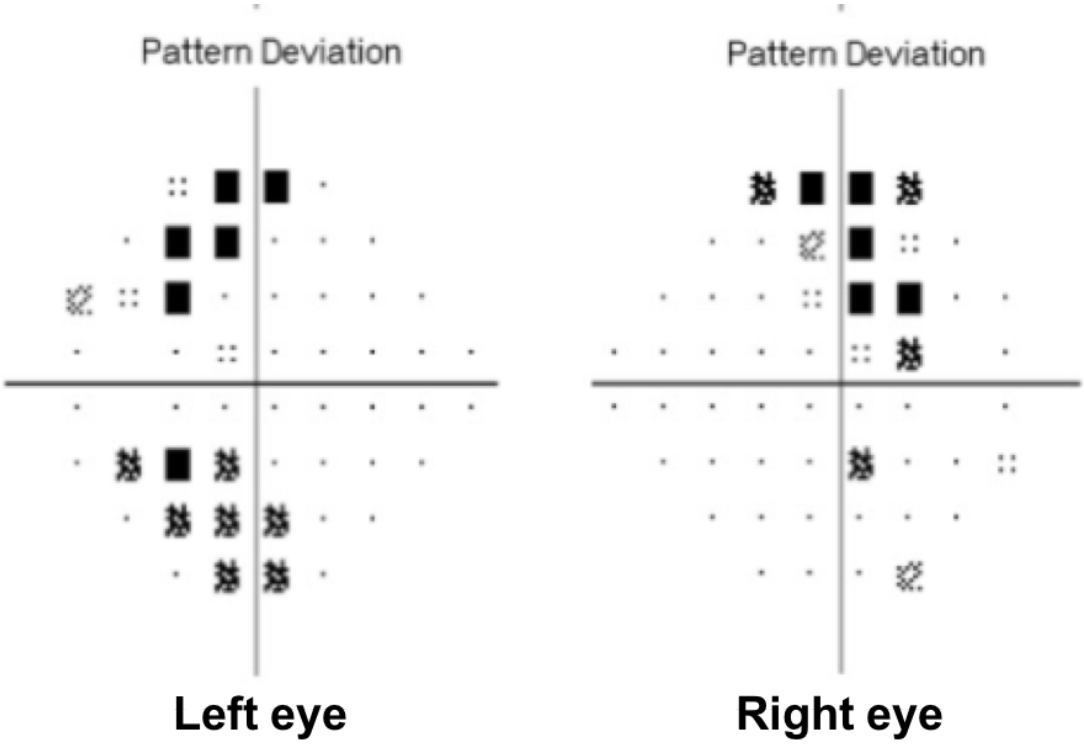

1. Non-specific changes
2. Arcuate defects
3. Bitemporal hemianopia
4. Cecocentral scotomas

10. What visual field defect does this patient have? 3. Bitemporal hemianopia

This patient's visual field appears to respect the vertical meridian and is best appreciated on the pattern deviation. Although there are some depressed points in the nasal visual field, there is a clear predominance in the temporal hemifield in both eyes. This is highly suggestive of a bitemporal hemianopia.

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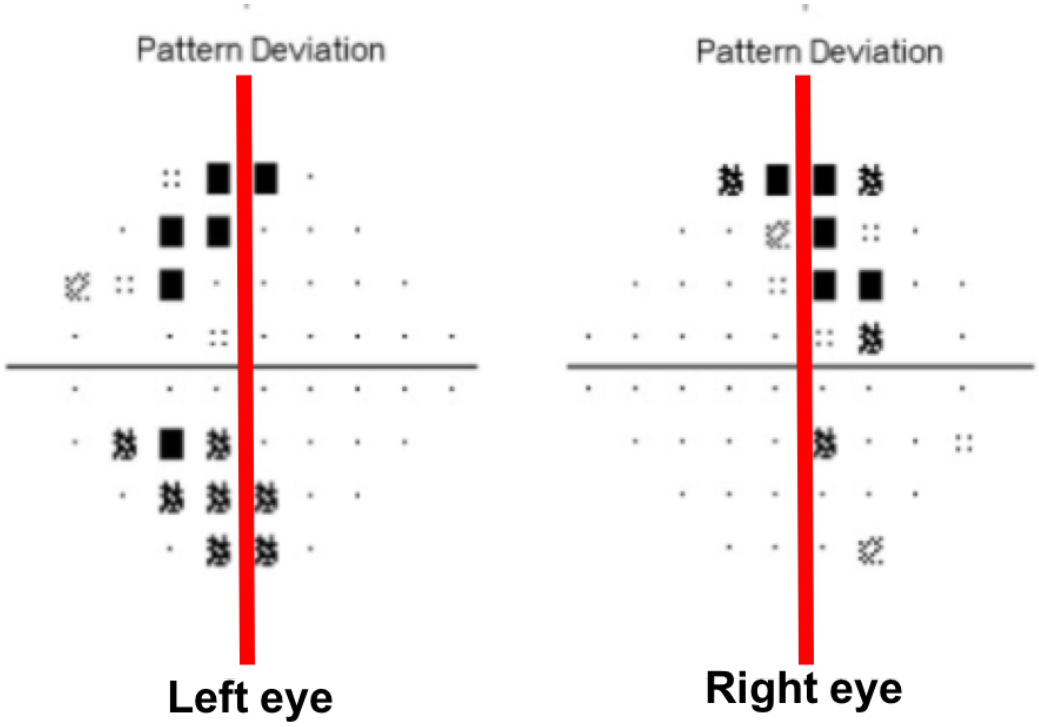

Although there are depressed points on both sides of the vertical meridian, there is a clear difference on the number on the temporal side of the visual field in both eyes (bitemporal hemianopia).

11. What is the next best course of action for this patient?

1. Initiation of topical glaucoma medication
2. Laser trabeculoplasty
3. MRI of the brain and orbits with contrast
4. CT scan of the head without contrast

11. What is the next best course of action for this patient? 3. MRI of the brain and orbits with contrast

The patient has a visual field defect that respects the vertical meridian and optic nerve cupping with pallor of the neuroretinal rim. This is concerning for a compressive lesion of the optic chiasm and an MRI of the brain and orbits is the best way to assess this. The MRI revealed a large pituitary adenoma (shown below).

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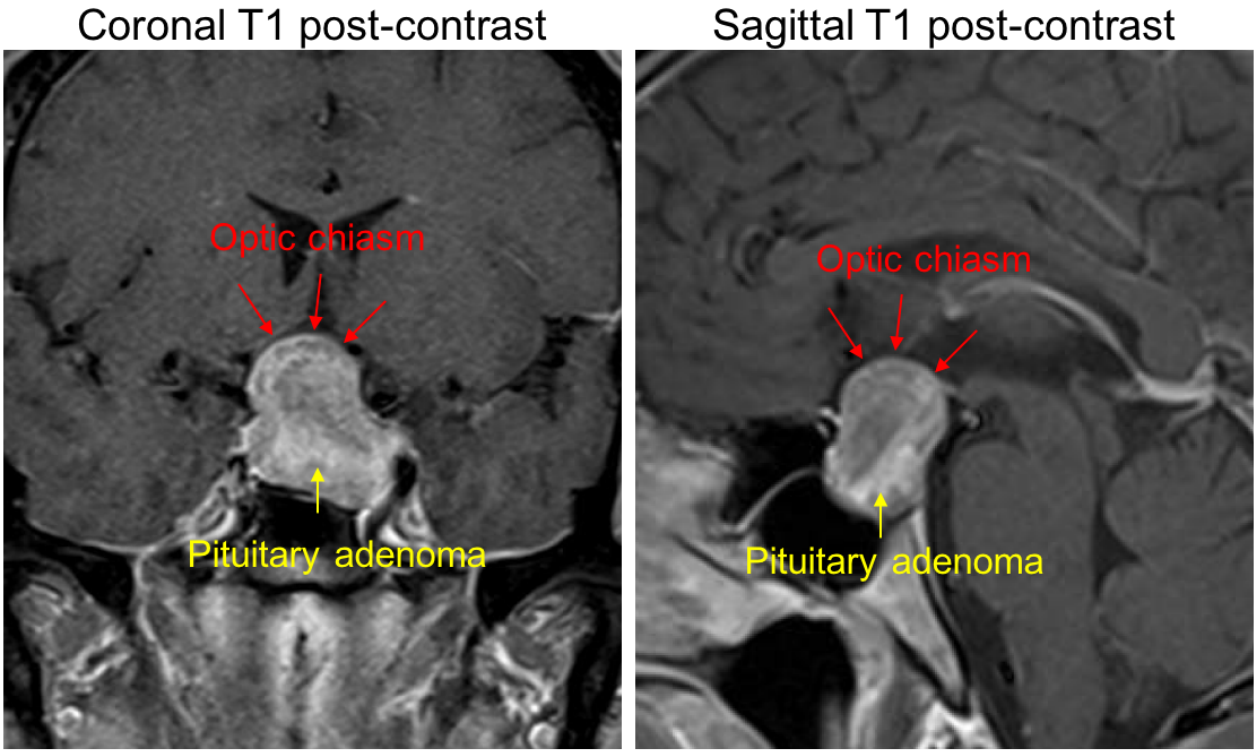

MRI of the brain and orbits with contrast demonstrates a large sellar/suprasellar mass most consistent with a pituitary adenoma. There is compression of the optic chiasm.

12. Which of the following is the best predictor of visual recovery after surgical removal of the pituitary adenoma?

1. Pre-operative visual acuity
2. Pre-operative retinal nerve fiber layer (RNFL) and ganglion cell complex thickness measured by optical coherence tomography (OCT)
3. Age
4. Duration of symptoms

12. Which of the following is the best predictor of visual recovery after surgical removal of the pituitary adenoma? 2. Pre-operative RNFL and ganglion cell complex thickness measured by OCT

Most visual recovery after resection of a pituitary adenoma occurs in the first 6-10 weeks, although there is continued improvement up to one year after surgery. Most patients will show improvement after surgery, but those with a normal OCT RNFL thickness are more likely to return to a near normal visual field. OCT of the RNFL and macular ganglion cell complex are recommended by the Congress of Neurological Surgeons as a prognostic tool in patients with visual loss from pituitary tumors

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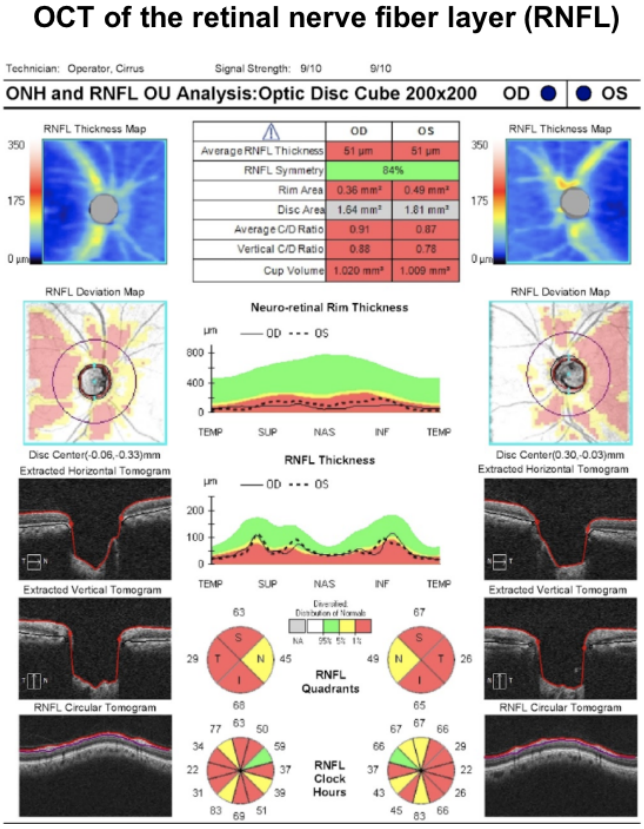
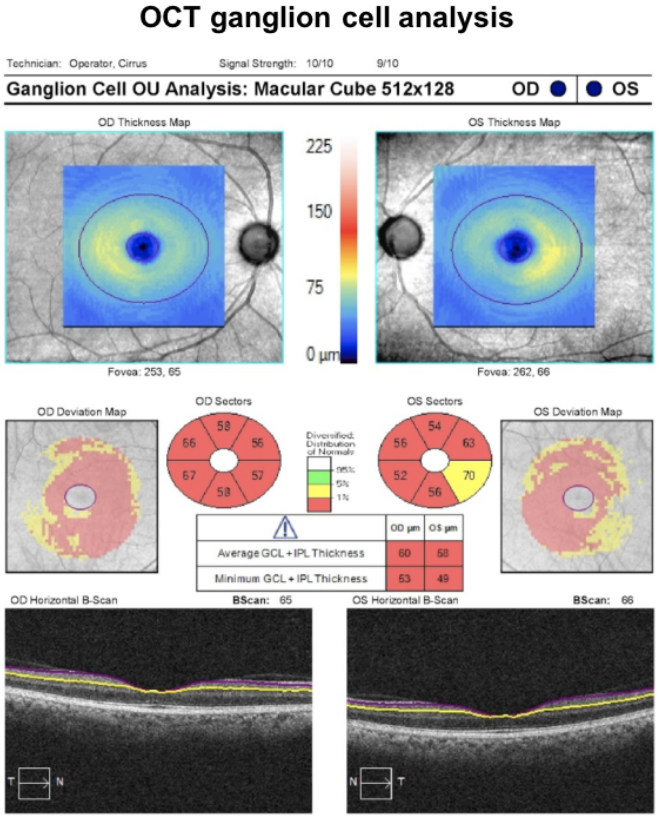
low RNFL thickness (51 um in both eyes), there is a lower chance of visual recovery compared to a patient with a normal RNFL thickness.





13. What else should be included in the pre-operative evaluation of this patient?

1. Extended cardiac monitoring
2. Genetic counselling
3. Endocrine evaluation for hypopituitarism
4. MRI of the spine

13. What else should be included in the pre-operative evaluation of this patient? 3. Endocrine evaluation for hypopituitarism

Endocrine evaluation to assess for hypopituitarism is recommended in all patients with pituitary adenomas. There is a high prevalence of hypopituitarism in patients with non-functioning pituitary adenomas and all anterior pituitary axes should be assessed. The cutoff values to initiate thyroid and adrenal replacement may be different in a patient with panhypopituitarism compared to isolated deficiencies and is best evaluated by an endocrinologist with expertise in this area.

Case Summary

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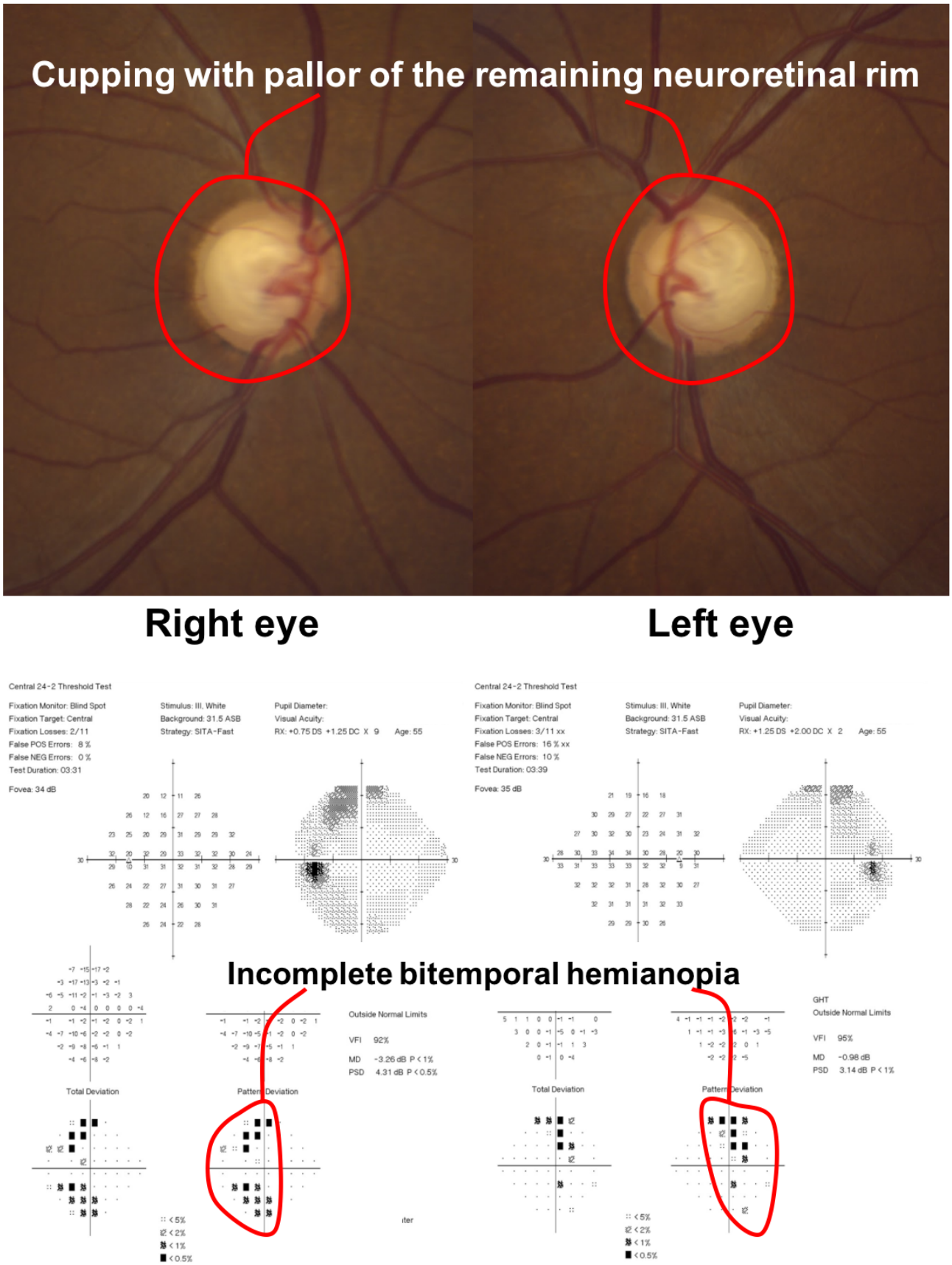
field showed an incomplete, bitemporal hemianopia. MRI of the brain showed a large sellar/suprasellar mass consistent with a pituitary adenoma. He had a transphenoidal resection of the tumor and had improvement in his visual field, although it did not completely return to normal.



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Further reading:

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